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CASE REPORT

Mc Cune-Albright syndrome with gigantism and hyperprolactinemia

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ABSTRACT. We describe the case of a 38-year-old man with typical Mc Cune-Albright syndrome and the unusual combination of both growth-hormone and prolactin hypersecretion. The patient was extremely tall, which is unusual in Mc Cune-Albright syndrome, suggesting that he did not have precocious fusion of the epiphysis, a common finding in this syndrome. Unfortunately the patient refused any treatment for his disease. A similar case has been previously described only in a 14-year-old boy.

INTRODUCTION

Mc Cune-Albright syndrome is a disease of unknown etiology consisting of polyostotic fibrous dysplasia of bone associated with brown pigmented areas of the skin and several endocrine dysfunctions (1, 2). These include sexual precocity, especially in females (3, 4), goiter and/or hyperthyroidism (4, 5), growth hormone (GH) hypersecretion (6), Cushing' disease (7) and accelerated skeletal growth (8). Recently the first case of Mc Cune-Albright with hypersecretion of both GH and prolactin (PRL) has been described in a boy (9). A hypersecretion of hypothalamic releasing factors has been suggested, but not proven, as the possible explanation for the endocrine manifestations of this syndrome (10).

We now report the second case of typical Mc Cune-Albright with concomitant GH and PRL hypersecretion in an adult man.

CASE REPORT

A 38-year-old man was referred to us by an otolaryngologist who had seen him because of bilateral ipoacusia and otorrhea. The patient was 203 cm tall and weighed 104 kg. He showed an asymmetric "leonine facies", had a severe kypho-scoliosis with normal hands and feet. Cutaneous "café au lait" spots from 1 to 10 cm were present on his back and left side of his neck and shoulder.

From the history it was possible to ascertain that the accelerated rate of growth and facial asymmetry were already present during childhood and probably even in infancy.

Radiological examination

A skeletal radiological survey showed an asymmetric overgrowth of the skull involving mainly the right side (jaw, mastoid and calvarium). At the vault the appearance simulated a long-standing cephalo-hematoma. The cranial bone structure appeared completely distorted with a mixed pattern of bone-forming and bone-destroying lesions (Fig. 1a and 1b). Because of such deformity any evaluation of the sella and petrous bones was not possible, furthermore the patient refused to perform a computerized assial tomography. The long bones, besides a generalized overgrowth, revealed diffuse cystic lesions mainly at the diaphyses of the right side (Fig. 2a, 2b, 2c). Moreover an enormous ballooning enlargement of the proximal end of the right ulna was evident (Fig. 3). The vertebral bodies, mainly at the dorsal tract, appeared increased in their antero-posterior diameters. There was also an exaggerated development of the spinous process (Fig. 4). The intervertebral spaces otherwise resulted normal. The flat bones presented an extensive overgrowth. Moreover radiolucent diffuse lesions were present at the pelvis and ribs.

Endocrine evaluation

As shown in Table 1, endocrinological studies revealed elevated basal levels of plasma GH (76 ng/ml) and PRL (234 ng/ml) which increased after TRH.

Thyroid hormones and basal TSH were normal with a low-normal response of TSH to TRH. Basal plasma cortisol, LH, FSH, PTH (c-terminal), calcitonin and testosterone were in the normal range. Serum calcium was normal and alkaline phosphatase was elevated (1210 U/L). Further assessment of hypothalamic-pituitary function, as well as any treatment were refused by the patient.

Key-words: Albright syndrome, polyostotic fibrous dysplasia, gigantism, hyperprolactinemia.

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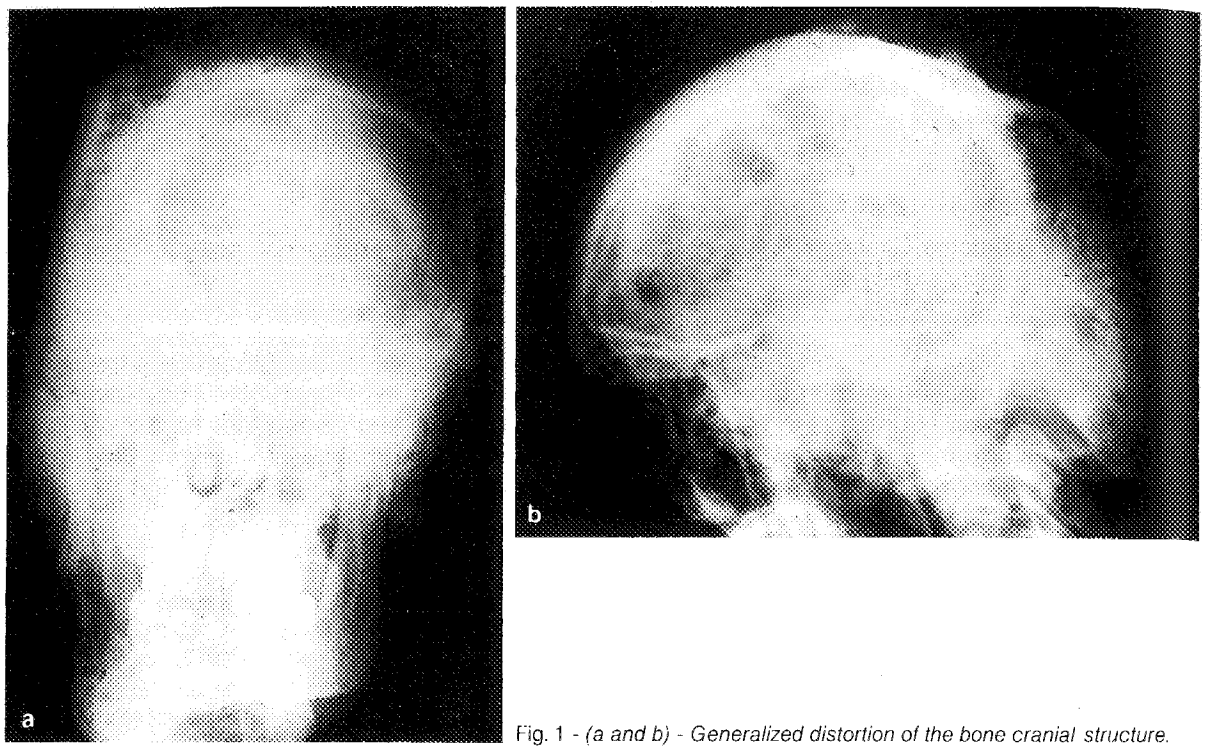

Fig. 1 - *(a and b) - Generalized distortion of the bone cranial structure.*

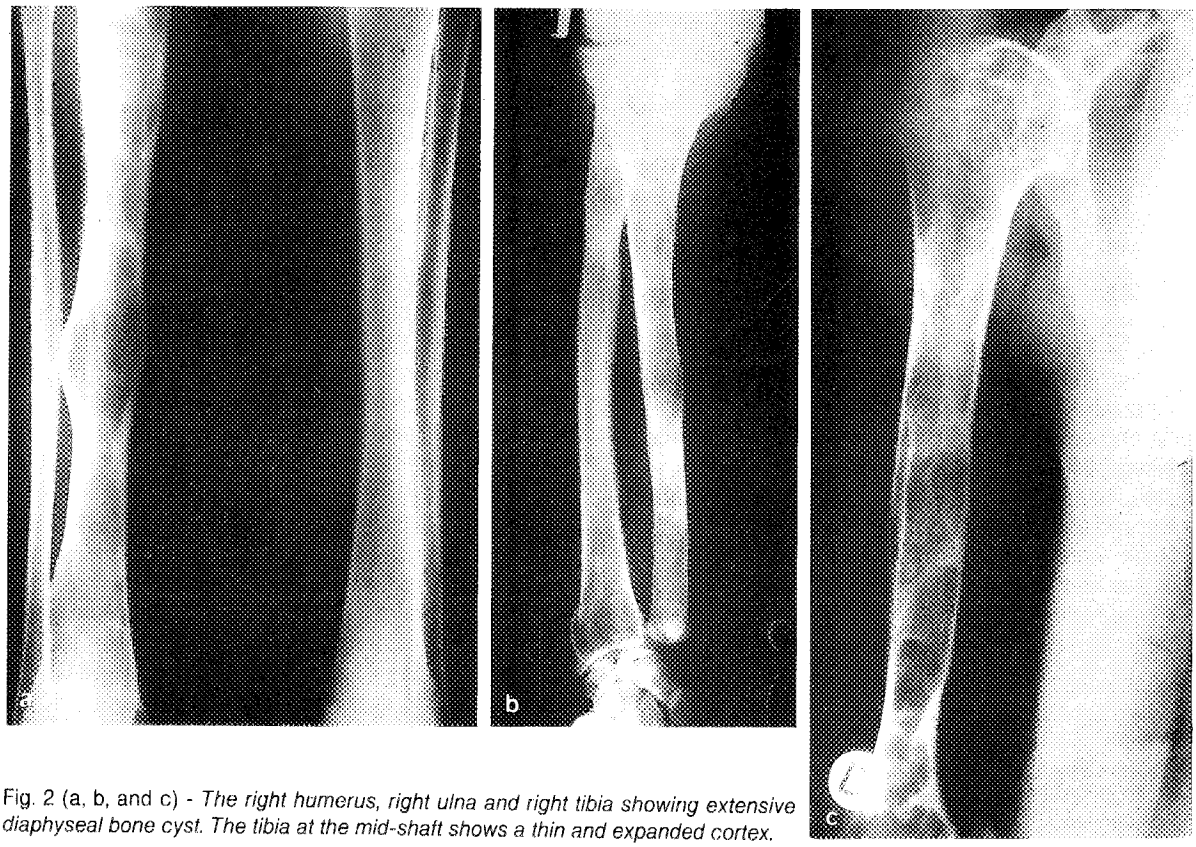

Fig. 2 (a, b, and c) - *The right humerus, right ulna and right tibia showing extensive diaphyseal bone cyst. The tibia at the mid-shaft shows a thin and expanded cortex.*

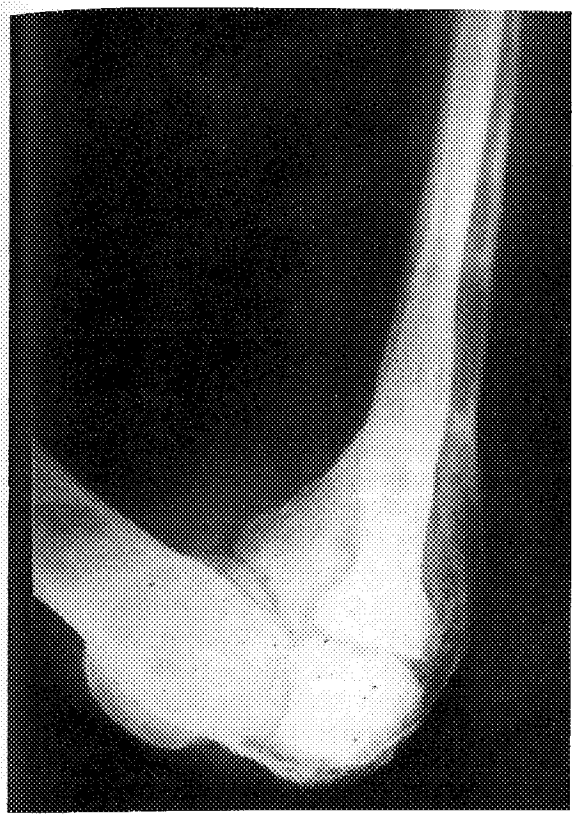

Fig. 3 - *Striking enlargment of the right ulna at the proximal epiphysis.*

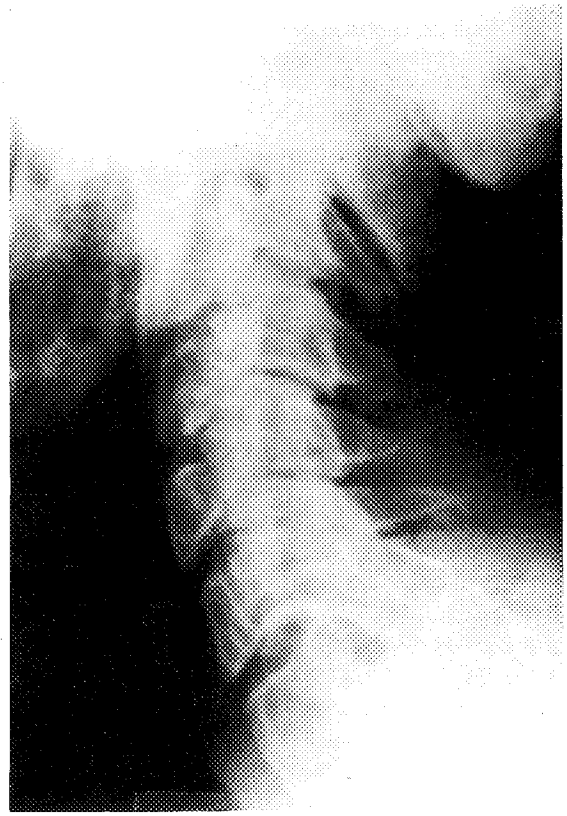

Fig. 4 - *Overgrowth of the spinous processes at C1-C6-C7.*

Table 1 - *TSH, GH and PRL after TRH test.*

	TSH μU/ml	GH ng/ml	PRL ng/ml
Basal	1.7	76	234
after TRH			
(200 μg) + 10'	4.9	90	216
+ 20'	7.5	104	280
+ 30'	6.3	108	265
+ 60'	6.7	88	260

DISCUSSION

All together the features described in this patient are consistent with a diagnosis of Mc Cune-Albright syndrome associated with gigantism and hyperprolactinemia. However some features of this patient are quite unusual and deserve comments. First, the patient was very tall, while patients with Mc Cune-Albright tend to have low height as a result of accelerated growth and precocious fusion of the epiphyses, even in the absence of sexual precocity. The clinical features in this patient began during childhood or even before, and at about age 10 facial deformities and high tall were already present.

It was not possible to determine whether the patient had sexual precocity, but in view of his height, this possibility seems unlikely, since it would have caused a precocious epiphyseal fusion with resultant low height. Probably GH hypersecretion during childhood was essential in determining his high tall.

Unusual is also the association of hypersecretion of GH and PRL. In Mc Cune-Albright such an association has been reported only in a 14-year-old boy whose endocrine alterations were suspected to be due to an autonomous pituitary adenoma (9).

Unfortunately our patient refused further morphological studies of his pituitary as well as further function tests. However the high levels of both GH and PRL and the paradoxical response of GH to TRH suggest the presence of an autonomous adenoma also in our case (11).

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